Sickle Cell Disease:

The Teacher Can Make A Difference

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Acknowledgments

This book is dedicated to children with Sickle Cell Disease, their families and their teachers.

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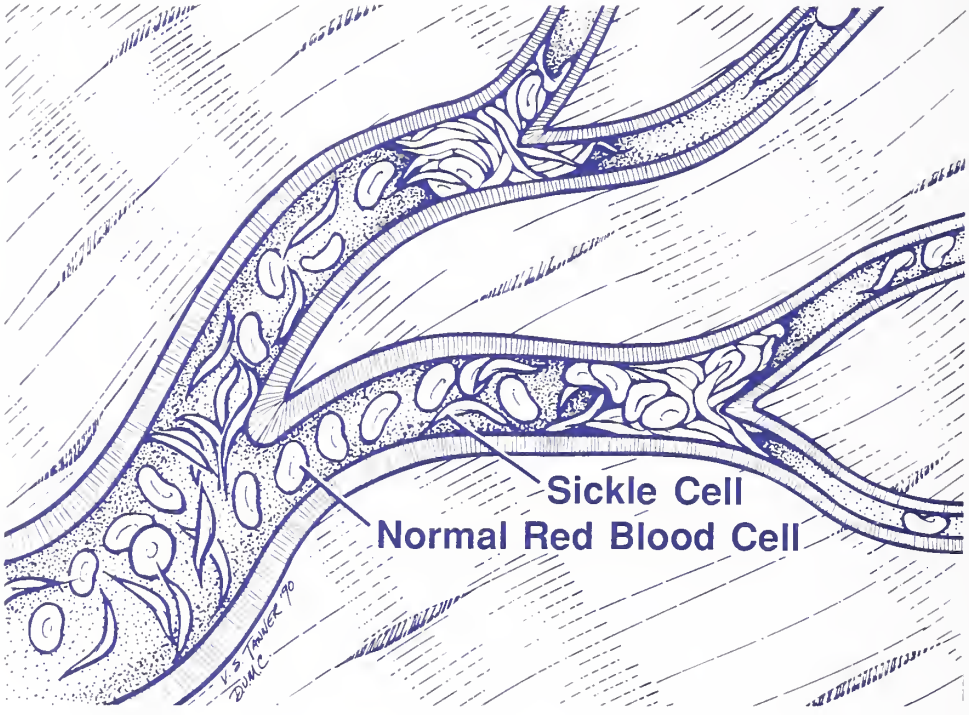
Sickle Cell
Normal Red Blood Cell

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Introduction

Teachers play a pivotal role in the formation of the educational foundation of our youth. Their role is critical in the child's intellectual and social development, as well as the development of the child's self-esteem and sense of self-worth.

A child's primary occupation is school, with the average child spending approximately thirty-five hours per week at school. Through repetitive encounters with children, teachers are able to reinforce ideas and information.

A number of factors may threaten the learning process. Chronic illness is one such factor. The child with a chronic illness can present a special challenge to the teacher. Frequent absences may lead to social isolation and academic failure, making it more difficult for the student to stay in the mainstream.

This booklet is designed to inform teachers about sickle cell disease (SCD) and to enable them to recognize the medical and psychosocial complications that may interfere with learning. Emphasis is placed on practical suggestions for specific interventions.

The booklet is divided into three sections. In the first section, some general information about sickle cell disease is presented. This is followed by a section on common medical problems. Each medical problem is summarized with a brief "RECAP," a list of "TEACHER OBSERVATIONS," and a list of "TEACHER INTERVENTIONS." The booklet concludes with a section on psychosocial aspects of sickle cell disease.

Definition of Hemoglobin

Hemoglobin is a protein in the red blood cells that transports oxygen throughout the body. As blood flows through the lungs, the hemoglobin in the red blood cells picks up oxygen and carries it to body tissues where it is released. Normal hemoglobin is called HEMOGLOBIN A (Hb A). SICKLE HEMOGLOBIN (Hb S) has a different chemical structure than Hb A. This difference in chemical structure causes the symptoms associated with SCD.

Definition of Sickle Cell Disease

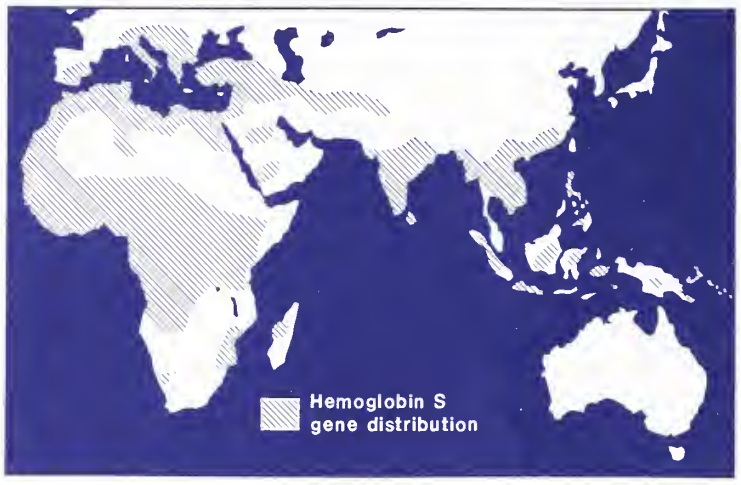
Sickle Cell Disease (SCD) is a term used to describe a GROUP OF INHERITED BLOOD DISORDERS characterized by a predominance of Hb S. The principal manifestations of the disease are life-long anemia and variable degrees of organ damage produced by the blockage of blood flow. The more common types of SCD include Sickle Cell Anemia and Hemoglobin SC Disease. In the most common form of SCD, Sickle Cell Anemia, almost all of the person's hemoglobin is S. In Hemoglobin SC Disease, the affected person has two abnormal hemoglobins, S and C.

Incidence of Sickle Cell Anemia

Approximately 1 in 400 African Americans is born with Sickle Cell Anemia. Sickle Cell Anemia affects many more Americans than inherited diseases such as Cystic Fibrosis, Phenylketonuria (PKU), and Hemophilia. It also has a higher incidence than Muscular Dystrophy and Tay-Sachs Disease.

Origin of Sickle Hemoglobin

The origins of Hb S can be traced to countries where malaria was common. The Hb S gene is found in people of African, Arabian, Iranian, Italian, Indian, Turkish, and Greek ancestry.



The Sickle Cell

Red blood cells in persons with normal hemoglobin are shaped like a biconcave disk and are flexible so that they can squeeze through the tiny capillary network in the body. Sickle cells are red blood cells that contain a predominance of Hb S and, though capable of transporting oxygen efficiently, undergo a shape change when oxygen is released to the tissues. The shape that they assume often resembles a sickle or crescent. Initially these cells can resume a biconcave disk shape when reoxygentated in the lung, but eventually they become damaged and are "stuck" in a sickled shape. These "irreversibly sickled" cells are rigid and can no longer easily travel through the capillary network. Blockage of blood vessels by the sickle shaped cells produces symptoms. The types of symptoms are related to the site where the vessels are blocked.

Anemia and Sickle Cell Disease

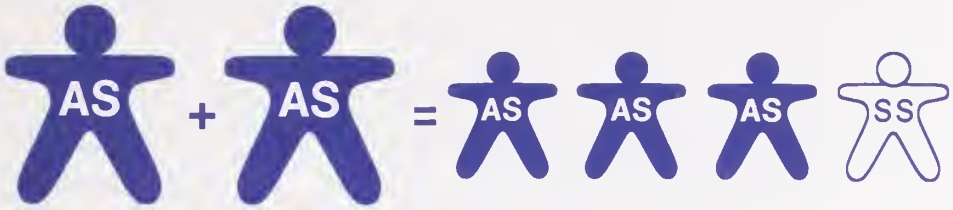
Anemia is a term indicating a reduced number of red blood cells. There are many causes for anemia. One cause is SCD. The sickling process leads to early destruction of the red blood cells. Normal red blood cells live 120 days but sickle cells last only about 12 days. Because the bone marrow, the site of production for all blood cells, cannot produce red cells fast enough to compensate for the reduced life span, the person becomes anemic. Anemia is a life long condition for persons with SCD.

Sickle Cell Trait

Sickle Cell Trait IS NOT A DISEASE. Individuals with Sickle Cell Trait have two hemoglobins in their red blood cells, Hb A and Hb S. These individuals inherit

Hb A from one parent and Hb S from the other. The dominant hemoglobin is Hb A. Under normal conditions, the red cells from persons with Sickle Cell Trait do not assume a sickled shape. Sickle Cell Trait NEVER changes to Sickle Cell Disease.

Approximately 1 in 10 African Americans has Sickle Cell Trait. When both parents have Sickle Cell Trait, there is a one in four chance with each pregnancy that the child will be affected with Sickle Cell Anemia, having inherited sickle hemoglobin from each parent.



Major Medical Manifestions of Sickle Cell Disease

Painful Episodes

Painful episodes are the hallmark of SCD. Pain is the most common re-curring feature of the illness. It is due to localized circulatory problems that arise when red blood cells sickle, stop blood flow, and deprive tissue of oxygen. The painful event is often preceded by infection, dehydration, or chemical imbalance in the body fluids.

Pain in SCD usually has a gradual onset. It is frequently described as an excruciating, gnawing pain. It usually occurs in the extremities and back but can occur anywhere in the body. The pain can be waxing and waning and can last from hours to over a week. Often it can be treated at home with oral medicines, but treatment may require a day-long clinic visit, overnight emergency room stay, or hospitalization.

Sometimes an "over the counter" pain reliever, such as Tylenol, can control the pain. It is common, however, for an individual to require an oral narcotic, such as codeine, to reduce the pain. Injections of stronger narcotics, such as Demerol or morphine, may be required.

A common pitfall in the treatment of pain is giving too little medication for fear of causing drug dependency. Addiction is a very uncommon problem in children with SCD. If drug seeking behavior is suspected, however, teachers should alert parents and the medical care team.

The painful episode does not always keep the child from school. With adequate medication, the child may be able to function in the school setting. If the child is attending school during a painful episode, the teacher should encourage the child to drink fluids. Recommended fluids include water, ice tea and soft drinks.

The child may have to stay home during a painful event. The simplest writing task can become impossible when the child's hand or arm is throbbing with pain. Frequent absenteeism may cause the child to fall behind or even repeat a grade. Teachers need to make provisions for these absences.

RECAP

- hallmark of disease
- most frequent recurring event
- pain medication usually required every 4-6 hours

TEACHER OBSERVATIONS

- child may appear frightened, upset, quiet, withdrawn
- child complains of pain, usually in legs, arms, back, chest and stomach
- child generally knows if pain is sickle cell related
- child may be able to stay in school with mild episodes of pain

TEACHER INTERVENTIONS

1. Try to recognize painful episodes early.
2. Demonstrate concern and support; comfort and reassure the student.
3. Alert parents but reassure them if child is having only mild pain. Decide with parents if child can stay in school.
4. Encourage fluids, especially after physical activities.
5. Send to school nurse, if available, to evaluate and give pain medication as needed.
6. Know how the child is expected to manage a mild painful event at school. The parent and physician can provide this information.
7. Encourage child to continue classroom participation if the pain is tolerable.

Worsening Anemia

Anemia is defined as a hemoglobin concentration below normal. The normal hemoglobin concentration for a healthy child is between 12 and 16 grams per deciliter (gms/dl), depending on age and sex. In Sickle Cell Anemia the hemoglobin concentration is usually between 6 and 10 gms/dl. With this degree of anemia, children have a decreased tolerance for vigorous physical activity. *Most children with SCD, however, can run, jump, swim, ride bikes and participate in physical education activities.* Children know what they can tolerate. We should trust their assessment of their physical limitations.

Children with Sickle Cell Anemia are at risk for suddenly developing worsening anemia. Although a rare complication, it may be serious enough to require a blood transfusion. It is often related to a viral infection which causes a temporary cessation of red blood cell production.

Even rarer is a condition known as "acute splenic sequestration." During this complication, which usually occurs in preschoolers, the spleen sequesters or traps the sickled cells resulting in a life-threatening situation of acute worsening anemia. Emergency treatment may be required to save the child's life.

RECAP

- anemia refers to a condition in which hemoglobin level is below normal

- people with anemia tire easily and should rest after strenuous physical activity
- sudden worsening anemia may require emergency care

TEACHER OBSERVATIONS

- child looks pale
- low tolerance for vigorous physical activity
- symptoms may worsen suddenly

TEACHER INTERVENTIONS

1. Encourage the child to participate up to his level of tolerance.
2. Allow recovery time after vigorous physical activity.
3. Inform physical education instructor of limitations.
4. If active participation not realistic, make student your "assistant," "scorekeeper," or "umpire."
5. Try to schedule more difficult classroom tasks in the morning when the child is more rested.
6. If child looks paler than usual, immediately inform parent.

Susceptibility to Infections

The most common sign of infection is fever, which cannot be taken lightly. Infection is the leading cause of death in young children with SCD. Fever deserves prompt evaluation. Parents need to be notified as soon as possible so that adequate medical attention can be sought. The child may need to be admitted to the hospital for intravenous antibiotics or may be treated at home with oral medicines if the infection is mild.

Pneumonia is one of the most common reasons to hospitalize children with SCD. Pneumonia is 100 times more common among children with SCD than among children in the general population. The most frequent symptoms of pneumonia are fever, cough, and increased respiratory rate.

RECAP

- fever is a cardinal feature of infection
- infections are a major problem in SCD
- infections account for a large proportion of hospitalizations
- infections are a major cause of death in preschool children

TEACHER OBSERVATIONS

- fever is a sign of infection
- cough and rapid respirations may be due to pneumonia

TEACHER INTERVENTIONS

1. If fever is suspected, inform school nurse and/or parents promptly. Take temperature if possible.
2. If parents cannot be reached, call child's health care provider for advice.
3. Be prepared with alternate telephone contacts in case parents cannot be reached.

Gallstones

With the rapid destruction of red blood cells in SCD, large amounts of hemoglobin are released. The hemoglobin molecules are further broken down, forming an excessive amount of bilirubin. This pigment accumulates in the gallbladder and contributes to the formation of gallstones. The gallstones can cause numerous problems, including nausea, vomiting, and abdominal pain. When these symptoms appear, and are attributable to gallstones, surgery is performed to remove the gallbladder. This complication is a relatively common problem in school-age children with SCD.

Children with SCD need to be transfused with normal red blood cells prior to surgery to reduce the risks associated with general anesthesia. Gallbladder surgery is often elective and can be planned in advance. Preoperative transfusions, hospitalization for surgery, and postoperative recovery will necessitate an extended absence from school.

RECAP

- many school-age children with SCD will develop gallstones
- gallstones can be serious; surgery may be required
- may cause an extended absence from school

TEACHER OBSERVATIONS

- symptoms of nausea, right upper abdominal pain, shoulder pain, and vomiting may be due to gallstones
- symptoms of gallstones may be associated with eating
- increased levels of bilirubin will cause the child to appear jaundiced (white part of eyes has yellowish tint)
- classmates may tease child with "yellow eyes"
- if required, surgery is usually elective and can be planned
- child likely to miss 1-2 weeks of school to recover from surgery

TEACHER INTERVENTIONS

1. Alert parents of symptoms and make student as comfortable as possible.
2. Explain to other children the reason for the absenteeism and "yellow" eyes.
3. Provide lesson plans for days missed for transfusions, hospitalization, and recovery from surgery.
4. Homebound instruction or special tutoring may be required as recovery from abdominal surgery may extend 1-2 weeks beyond hospitalization.

Delayed Growth and Sexual Maturity

On the average, puberty in children with SCD is delayed about two years compared to normal children. Delayed growth and pubescence can undermine self-esteem. Small size and sexual immaturity may be a focal point of concern for the teenager with a heightened interest in body image.

RECAP

- growth and sexual maturation may be slower than peers
- may be emotionally disturbing during adolescence

TEACHER OBSERVATIONS

- child may appear years younger than age
- small stature may prompt peers and teachers to regard child as younger than actual age
- teasing by classmates
- exclusion from group activities
- poor self-esteem

TEACHER INTERVENTIONS

1. Educate classmates. The topic of SCD could be used for a health education project. Attention to the visible signs of the disease could be a focus of discussion.
2. Treat the child appropriately for age.

Stroke

Stroke occurs in a small percentage of children with SCD. If a child has a stroke, an individualized educational program should be developed and can be facilitated by psychometric testing at one of the state funded Developmental Evaluation Centers. Physical therapy and speech therapy are often required.

If a stroke occurs, red blood cell transfusions are administered monthly in an effort to reduce the risk of recurrent stroke. This treatment lasts for years and ultimately requires the concomitant use of a medicine to remove excessive amounts of iron which accumulate as a consequence of the blood transfusions.

Symptoms of stroke include difficulty with speech and vision, confusion, paralysis, spasticity, seizures, and muscle weakness. The child who suffers a stroke may make a total recovery, but usually there is some residual deficit.

RECAP

- occurs in 5-10% of children with Sickle Cell Anemia
- can be a devastating complication
- can leave student mentally and physically handicapped
- long term transfusion therapy required after stroke

TEACHER OBSERVATIONS

- visual disturbances, headaches, paralysis, unusual weakness, imbalance, clumsiness, seizures, and loss of speech may be signs of stroke

TEACHER INTERVENTIONS

1. If signs of stroke are observed, emergency medical treatment is required. Alert parents or other designated emergency contacts. Notify school nurse if on site.
2. If a child has a stroke, tailor educational program; obtain psychometric testing to define the child's abilities.
3. Report changing performance patterns to parents.
4. Teacher may opt to provide class assignments on days child is scheduled for transfusion. There is usually time to work on assignments during the transfusion therapy.

Hip Problems

Lack of adequate blood flow to the hip can cause some of the bone to die, producing severe pain and limitation of motion of the hip joint. This condition is known as aseptic necrosis of the hip. Bracing prevents dislocation of the hip and can help relieve pain. The brace is a cumbersome appliance, however, and can have an overwhelming psychological impact on the school-age child or adolescent.

RECAP

- bone and joint problems can be debilitating
- a cumbersome brace may be required

TEACHER OBSERVATIONS

- change in student's gait should be reported
- pain in hip joint may be due to bone destruction

TEACHER INTERVENTIONS

1. If a brace is required, it may be necessary to rearrange classroom assignments to facilitate accessibility. Seating assignments also may need to be changed.
2. Be sensitive to psychological impact that gait disturbance or bracing may have on the child.
3. The child wearing a brace should not be excluded from school activities (e.g., Child should participate in physical education. Make student an assistant instructor, scorekeeper or umpire).

Susceptibility to Dehydration

SCD can damage the kidney tissues, leading to changes in the kidneys that do not allow them to concentrate the urine. The kidneys of individuals with SCD do not retain water effectively. This results in large volumes of urine and an increased risk of becoming dehydrated. Dehydration may trigger a painful episode.

RECAP

- kidneys do not conserve fluids
- large amounts of urine are excreted
- dehydration may trigger a painful episode

TEACHER OBSERVATIONS

- frequent requests to use the toilet
- frequent requests for a drink of water

TEACHER INTERVENTIONS

1. Encourage frequent fluid consumption, especially on hot days or when physical activities are strenuous.
2. Permit frequent trips to use toilet.
3. Inform classmates about these special needs.

Psychosocial Issues

SCD may adversely affect the physical and psychosocial well-being of the child. These adverse effects are intensified by the lack of adequate information, counseling, and social supports. Knowledge of medical aspects of SCD and its psychosocial impact enhances the teacher's ability to relate to the affected child in a supportive manner, provide the child with accurate information and foster the child's self-esteem.

Compared to their healthy peers, children with SCD experience additional physical and psychosocial stresses . The physical stresses include those produced by recurrent pain, hospitalizations, and uncomfortable therapeutic procedures. Emotional stresses include those produced by school absences, and feelings of anxiety, inferiority and hopelessness. In addition, the older child may feel guilty because of the financial pressures placed on the family as a result of costs associated with treatment of the illness.

Teachers and other school personnel should not make general assumptions about the medical or psychosocial status of children with SCD. Assumptions made without substantiating information may reinforce sick behavior and poor self-esteem. While many children with SCD are well adapted emotionally to handle the stresses produced by their illness, others are not. Some children with SCD enter school from an environment in which they have been "over protected" as a result of their illness. These children inadvertently have been taught to assume a "Sick Role." The "Sick Role Syndrome" is characterized by over dependency, helplessness, self-pity, excessive and unpredictable levels of frustration, reluctance to try new tasks, negative attitudes and poor self-image. This syndrome

results in dysfunctional behavior that inhibits participation in age appropriate developmental activities and impedes both social development and the acquisition of a quality education.

The teacher should evaluate the child's needs and develop a comprehensive individualized education plan. This approach is often best developed in concert with the parents and the child's health care providers.

The teacher needs to be comfortable talking about SCD. A classroom climate where the student feels comfortable enough to participate in all activities will minimize tendencies toward negative attitudes and behaviors. Realistic expectations should be maintained in relation to the child's situation.

Creating opportunities for the child to exhibit independence is very important when working with children with SCD. Some children are overly dependent and resist taking responsibility. Other children may regress to dependency after a painful event or hospitalization and may become discouraged due to a lack of control over their situation. By creating opportunities for independence, the teacher can foster a child's coping abilities and increase the child's self-esteem.

Emphasizing progress is another positive intervention strategy. The life experience of many children has reinforced only the debilitating effects of the disease. Without the reinforcement of positive assets and accomplishments, children can become discouraged about their progress and their future. By listening and providing opportunities for the student to ventilate, the teacher can help the student recognize progress, which may lead to increased self confidence and self-esteem.

The teacher can help minimize the frustration associated with school absences by giving assignments well in advance of the due date and by scheduling deadlines for missed work to be completed. Also, the teacher can advocate for tutorial services as the need arises and inform parents and pupils of resources available within the school system.

Families need to know that teachers welcome their involvement. If parents or guardians feel unwelcomed or excluded from the educational process, it may be more difficult for them to be supportive and they may be less motivated to accept their roles as collaborative partners in the educational program. The teacher should consistently support the child and family by discussing current issues of concern and giving family members information on the student's progress.

Occasionally, counseling may be necessary to help the child cope with emotional stresses produced by the disease. In some instances, counseling may be needed for the entire family. The teacher, in conjunction with other school personnel, should be aware of appropriate resources for referral of children and families.

In the remaining sections of this booklet, several specific areas related to psychosocial issues are discussed. These areas include Absenteeism, Career Guidance, Peer Support and Health Education. Specific suggestions in each of these areas are provided.

Absenteeism

- May be largest obstacle to school success for some children with SCD

- Basis for low academic skill levels
- More than 50 days of the school year may be missed
- Frequent absences of short duration may not allow teacher the option of homebound instruction
- Children with SCD fall under the category of "Other Health Impaired," qualifying them for special services
- Parents should work closely with teachers and tutors to monitor student progress in the completion of assignments. They can reinforce the necessity of making up work at home

TEACHER OBSERVATIONS

- disadvantaged compared to peers who have few absences
- not able to respond to verbal questioning about study material
- academic success more difficult to achieve with each school absence
- recurrent absenteeism may promote more expression of disease symptoms as a response to pressures of school work

TEACHER INTERVENTIONS

1. Plan in advance for lost time.
2. Establish early contact with parents and health care provider to determine child's status and current medical and psychosocial needs.
3. Recruit parental involvement in teaching process; encourage parents to work with child at home.
4. Provide parents a course outline that indicates when assignments are due.
5. Develop a mechanism for the exchange of materials and assignments between the teacher and the homebound student.
6. Expect that most make up work will be done after the child returns to school. Allow adequate time for this work to be completed.
7. Schedule tutoring sessions.
8. Recruit capable peers as tutors.
9. Understand the reason for absence. Request doctor's note for extended absences.
10. Support the child's endurance of mild painful episodes when child is attending school.
11. Provide the parents with specific, concrete hints on how to help child at home with assignments.
12. Expect absences, but do not excuse them. Focus on make up.
13. ENCOURAGE STUDENT.

Vocational Counseling

- Sedentary occupation as an adult may be necessary
- Focus should be on potential for productive activity

TEACHER OBSERVATIONS

- physical limitations may impact on employment opportunities
- expectations may be diminished

TEACHER INTERVENTIONS

1. Help the child or adolescent to understand physical limitations and encourage the selection of course work that will prepare the student for vocations that are realistic.
2. Refer to school counselor when additional support is needed.
3. Refer for vocational counseling when appropriate.
4. ENCOURAGE, ENCOURAGE, ENCOURAGE.

Peer Support

- Fosters a child's self-esteem
- Child may respond more readily when encouragement comes from peers

TEACHER OBSERVATIONS

- tendency toward isolation within the school setting
- last chosen for activities
- reluctance to participate in group activities and/or to volunteer for classroom activities
- lack of self-confidence

TEACHER INTERVENTIONS

1. Highlight child's positive characteristics so that classmates see child as competent in certain areas, thus elevating the child's self esteem.
2. Provide opportunities for peer tutoring.
3. Choose child for classroom jobs.
4. Assign leadership roles to child in classroom activities and projects.
5. Encourage participation in extracurricular activity in which the child should excel.
6. PRAISE, PRAISE, PRAISE.
7. ENCOURAGE, ENCOURAGE, ENCOURAGE.

Health Education Unit on Sickle Cell Disease

- The average student has limited information (if any) about SCD
- Students may have misinformation about SCD
- Child with SCD has special needs that are not obvious to other children in the class
- It may be useful to present SCD as a topic for a health education unit

TEACHER OBSERVATIONS

- student comments and questions reveal a level of misunderstanding about SCD
- information on SCD is not readily available
- resentment expressed about the "special favors" granted a child with SCD, e.g., toilet and water fountain privileges
- Teasing a child with SCD who moves "too slow" or tires out quickly

TEACHER INTERVENTIONS

1. Integrate educational presentations on SCD into the curriculum of several courses (e.g., biology, geography, health).
2. The nearest sickle cell center may be contacted to help with curriculum development, as well as selection of appropriate educational materials.
3. Contact the sickle cell center to identify local sickle cell programs and/or resources in the area.
4. The child's medical doctor, physician assistant, nurse, or social worker can be invited to address the class. Local Health Departments can also provide educational presentations that are age appropriate.
5. Ask the child affected with SCD to help present the material to the class. The child can give personal testimony regarding life experience with SCD.

Student Name: _____

Parent/Guardian Name: _____

Home Telephone: _____

Work Telephone: _____

Other Telephone: _____

Medical Contact Name: _____

Telephone: _____

NOTES

NOTES

For Additional Information Contact:

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